CONGENITAL MALFORMATION OF THE POSTERIOR URETHRA*

By Oswald S. Lowsley, M.D.

HOUSE SURGEON, BELLEVUE HOSPITAL, NEW YORK CITY

(From the Pathological Laboratory of Bellevue Hospital (Charles Norris, director), Department of Bellevue and Allied Hospitals, New York City.)

This report concerns a case of congenital obstruction to urinary outflow due to an anomaly in development at the outermost end of the verumontanum.

J. M., aged three and one-half months, was admitted to the department of pædiatrics at Bellevue Hospital on July 30, 1913, as an urgent case. He had a temperature of 105°, and died a few hours after admission with œdema of lungs. The only history obtainable was to the effect that the child had been ill for a few days only, and the mother had noticed nothing unusual about its micturition.

Pathological Report.—The points of interest in the necropsy refer almost entirely to the genito-urinary tract, so the complete description is omitted.

The abdomen is distended. There is a hydrocele of the right cord. The peritoneal cavity is free from fluid or adhesions. There is a rather large fluctuating mass in the region of each kidney, the ureters being markedly dilated and tortuous. The bladder is distended and feels firm. The left kidney shows fetal lobulations and is much larger than the right, measuring 8 cm. from the upper to the lower pole. It is 3.1 cm. in width and 3.5 cm. thick. The pelvis of the kidney is so dilated that it considerably exceeds the kidney itself in size, measuring 4.5 cm. in width and 5 cm. from the edge of the kidney proper to its farthermost edge. The kidney has the appearance of a cap placed on a membranous sac and is very definitely distinguishable from the latter. On section it is observed that the kidney tissue varies in thickness from 1 mm. to 5 mm. and is seen to be flattened out into a shell, no sign of pyramids remaining, and occasional depressions are the only distinguishing marks of the calyces. The tissue of the pelvis is thinned out until it is of tissue paper thickness; the greatest diameter of the inside of the kidney and pelvis is 7.5 cm. There is a slight constriction which marks the beginning of the ureter. This measures 2.2 cm. in circumference. The capsule, which is rather thin, strips easily. The left ureter is enormous in size and markedly increased in length, measuring 13 cm. It is very tortuous and irregular in size at various points throughout its course. The average circum-

*Read in the section on Genito-Urinary Surgery, New York Academy of Medicine, Wednesday, November 19, 1913.

ference is 3.8 cm. There is much folding, and the wall is, comparatively speaking, decidedly thin. The ureter passes obliquely through the bladder wall in the usual manner, and one is struck by its minuteness at this point, compared to the size of the structure elsewhere, because it can be penetrated only with a probe.

The right kidney shows fetal markings rather more pronounced than does the left. On the anterior and outer surfaces of the lower half of the organ, there are thirteen retention cysts, which vary in size from a pin head to 4 mm. in diameter. On the posterior surface at the junction of the middle and upper thirds, there is another small cyst which measures 6 mm. in diameter. The organ is 6.5 cm. in length, 2.8 cm. in width, and 2.6 cm. thick. The pelvis on this side is very similar in appearance to the one on the left, but is not proportionately so large. It measures 3.6 cm. from the kidney substance to the outermost portion of its circumference. It is 3 cm. in width. This organ, as does its fellow, bears a very close resemblance to a cap placed over a membranous sac which is the dilated pelvis.

On section the kidney cortex is found to vary in thickness from 1 mm. to 9 mm. The greater thickness at some portions is due to the fact that several prominent fetal lobulations are cut through. The entire anatomical arrangement of the organ is distorted, as in the left, the pyramids being entirely obliterated and an occasional depression marking the site of a previous calyx; a shell of kidney tissue of varying thickness is the impression one gets in examining the organ. On the outside of the thin pelvis are seen the veins and arteries coursing to the kidney proper. The greatest diameter of the inside of the urine-containing cavity is 4.7 cm. The beginning of the ureter is marked by a constriction and a slight folding of the mucosa. This measures 1.2 cm. in circumference, and is the smallest portion of the ureter above the bladder. On this side the ureter is much smaller, particularly in its upper half, where it averages 2 cm. in circumference. It becomes larger lower down, and at one point reaches a circumference of 3 cm. It is somewhat longer and more tortuous than its fellow, being 14 cm. in length. It shows the same extraordinary reduction in size as it enters the bladder.

Blood Supply.—Right kidney: An artery about 2 mm. in diameter is given off from the lateral surface of the aorta. It passes upward and backward, and before reaching the dilated kidney pelvis. it gives off a branch which divides into three smaller ones and supplies a part of the upper pole of the kidney itself. The main trunk passes to the dilated pelvis, and there divides, one smaller branch running posteriorly between the kidney substance and the pelvis, giving off branches to both, and finally enters the kidney substance at about midway between the two poles. The larger branch passes anteriorly and, dividing into several smaller branches, becomes lost in the substance of the upper pole. A second artery, similar in size, comes off the aorta 7 mm. below the one just described. It passes almost horizontally outward, attaches itself to the anterior surface of the pelvis, where it divides into two larger branches and a smaller branch which supply the pelvis, and passing outward give off branches to the lower one-half of the kidney substance itself.

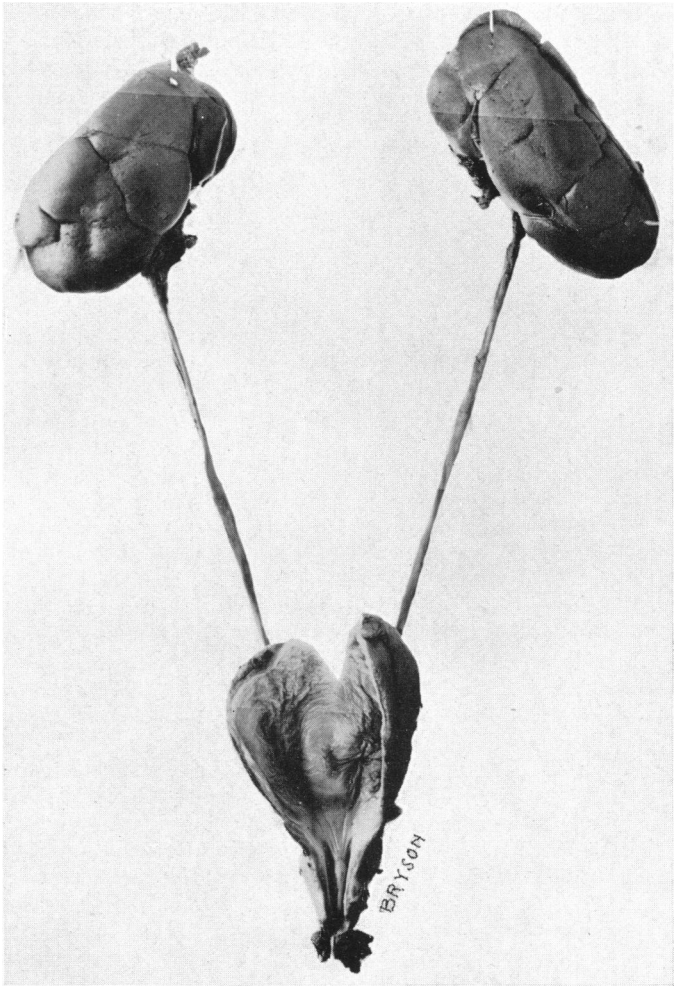

FIG. 1.—Normal bladder and kidneys of a 4-months-old boy (three-fourths natural size).

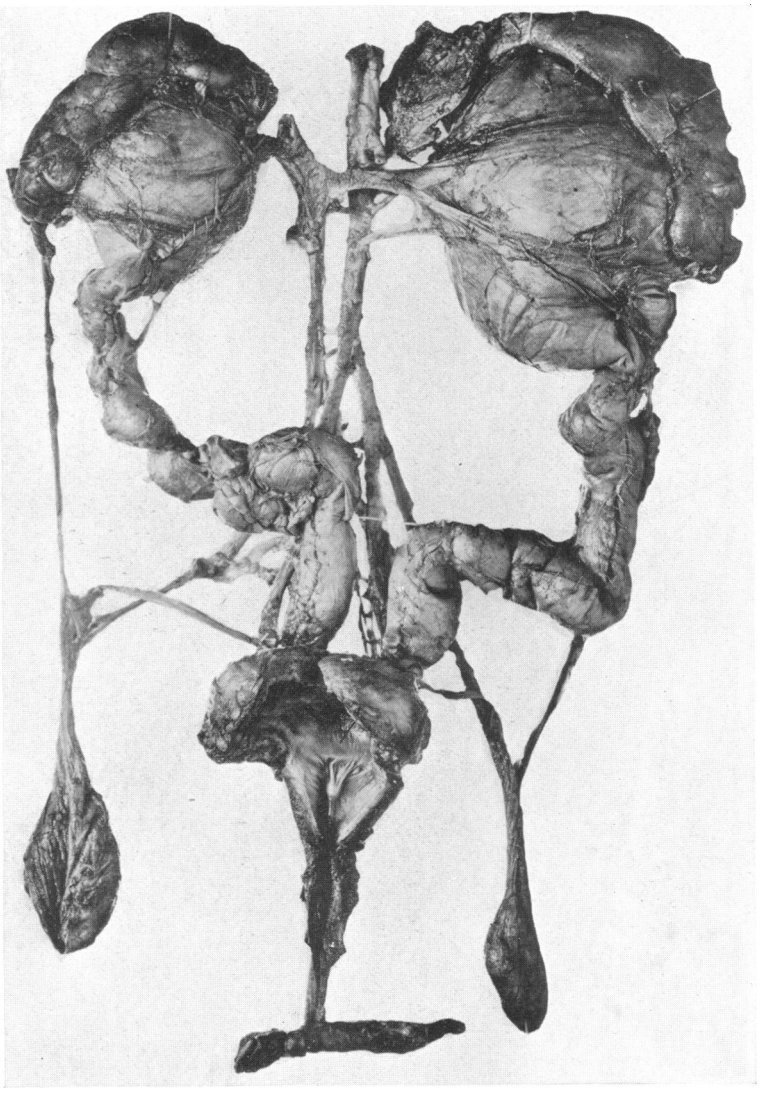

FIG. 2.—Anterior view of a case showing hydro-ureter and hydronephrosis, in a boy aged three and one-half months (one-half natural size).

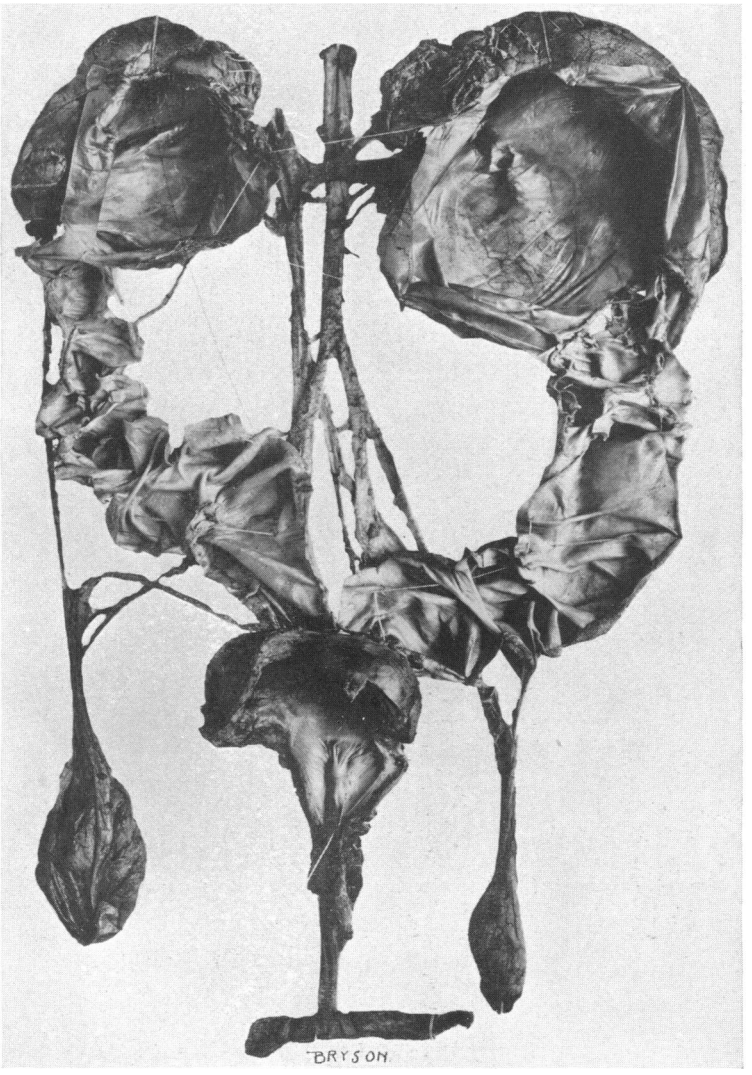

FIG. 3.—Same as Fig. 2, except that the ureters and kidney pelves are opened up and spread out, showing folds. There are probes in the ureters where they pass through the bladder wall. There is also a probe in the slit-like opening through the point of obstruction.

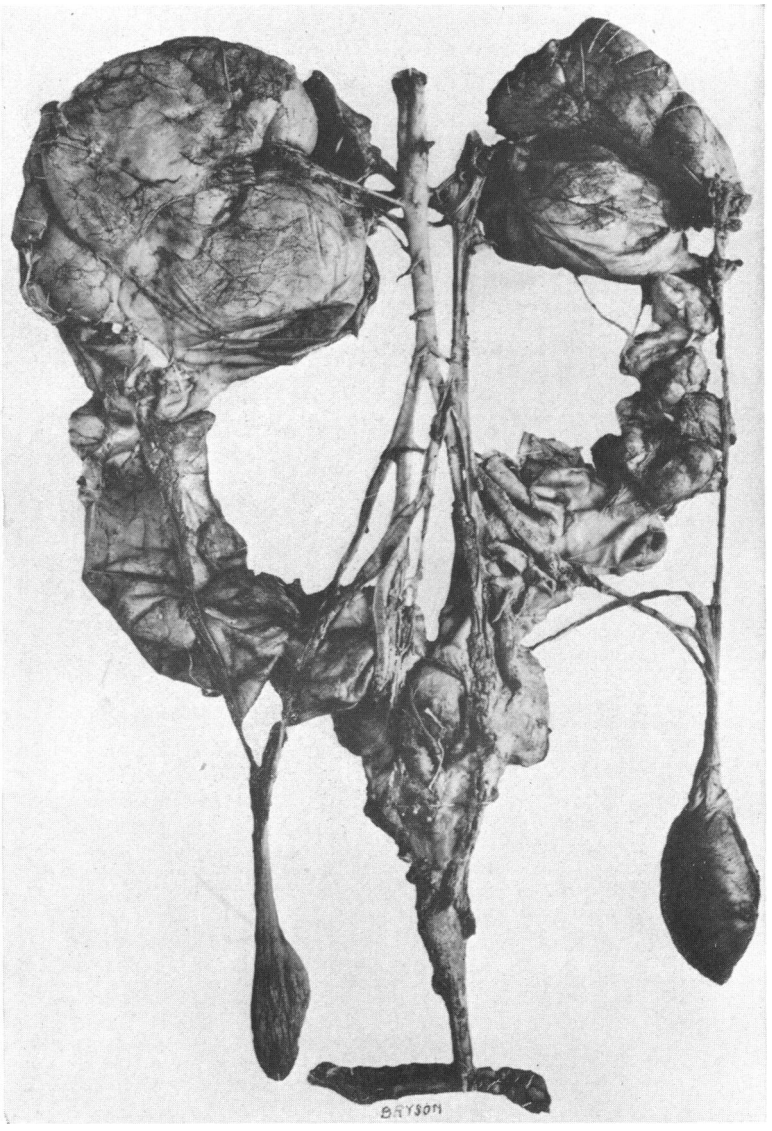

FIG. 4.—Posterior view, showing the left suprarenal gland and the arrangement of blood-vessels (one-half natural size).

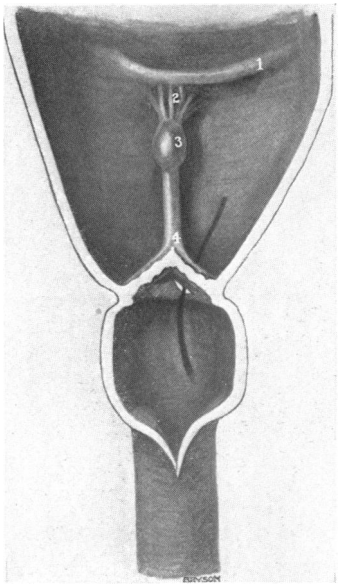

FIG. 5.—Diagram of posterior urethra. 1, lower end of internal (vesical) sphincter; 2, strands of tissue extending between trigonum vesicæ and upper end of verumontanum; 3, verumontanum; 4, lower end of verumontanum, showing the obstruction, with a small, slit-like opening on the floor to the left of the middle line. The urethra is laid open and shows a dilatation above and below the point of obstruction.

Veins accompanying this arterial supply, emptying into the ascending vena cava by four small trunks varying in size from 1 to 2 mm. in diameter.

Left kidney: The upper posterior half of the left kidney is supplied by an artery 1.75 mm. in diameter which arises from the lateral wall of the aorta about 3 mm. below the uppermost renal artery on the opposite side. This vessel passes outward until it becomes attached to the pelvis of the kidney 6 mm. from the kidney substance where it divides into two main branches, one of which passes down between the kidney substance and membranous pelvis, as far as the junction of the middle and lower thirds, and supplying both structures; the other passes directly to the upper pole where it subdivides and supplies this portion of the organ. A second larger artery, 2 mm. in diameter, is given off 4 mm. below, and considerably anterior to, the one just described. It passes outward to the pelvis and courses along its surface, finally dividing into branches which supply the anterior upper two-thirds of the kidney. A third artery, 1 mm. is diameter, passes horizontally outward, becomes attached to the pelvis and, traversing it, finally divides into branches which supply the lower one-third of the organ.

The veins of this organ are interesting, in that the blood of the entire structure is collected in vessels which accompany this unusual arterial supply, and unlike those on the opposite side, the veins here join at the anterior upper portion of the pelvis and empty into the ascending vena cava in one large trunk 1.7 cm. long after receiving a branch from the adrenal on this side.

The bladder seems to be about the usual size, but on section a most remarkable thickness of its walls is noted. This thickness is quite uniform over the entire organ and averages 8 mm. Throughout the entire fundus, there is noted a trabeculation with the formation of small cellules. The inter-ureteral ridge is present, but not marked, and the trigonum vesicæ is covered with a smooth mucosa, but is not particularly prominent. The ureteral orifices are slightly increased in size and their sphincters apparently obliterated. There is a ridge at the apex of the trigonum vesicæ which marks the site of the vesical sphincter. This latter structure is completely dilated so that the bladder and prostatic urethra are continuous and together form a funnel-shaped reservoir, the apex of which is found on a level with the apex of the prostate.

In the dilated posterior urethra are observed a number of prostatic duct openings. The verumontanum is peculiar in that six small bands are seen extending from the apex of the trigonum vesicæ to the largest portion of that structure where they seem to become imbedded in and form an integral part of it. At its widest portion, the verumontanum measures 3 mm. At this same point it is 3 mm. in height. At its lowermost portion, a particularly interesting arrangement is noticed. Ordinarily the verumontanum, which is formed by the ingrowth of the Müllerian and Wolffian ducts and their accompanying muscular coats, as described previously by the author,[1] becomes smaller and smaller at its lower portion where its fibres, about 1 cm. below its upper end in a specimen of this age, finally disappear by spreading out on the floor and

sides of the urethra in from 2 to 6 strands, some of which attach themselves to the walls. In this case some of the tissue of the verumontanum is disposed in the usual way, but a considerable portion of it continues down to the membranous urethra, where it divides into two portions and then attaches itself intimately to the entire urethral circumference with the exception of a very small slit-like opening on the floor of the urethra, just to the left of the median line which is lined with mucous membrane and could be penetrated with a fine probe.

The manner in which the division into two rather thick membranous bands occurs, and their attachment to the entire circumference of the urethra with the exception of a small aperture on the floor to the left of the median line, and also the fact that the entire structure is more or less dome-shaped, make the term "diaphragm," suggested by Dr. Hugh H. Young, seem most appropriate in referring to this anomaly. Almost complete obstruction to urinary outflow is caused by this unusual arrangement. Below the point of blocking, the bulbous urethra is considerably dilated.

There is a very large hydrocele of the right cord. Both testicles, the vasa deferentia, seminal vesicles, and the prostate are normal in appearance.

Microscopic Description.—Kidney: The section is a typical picture of compression of the kidney due to hydronephrosis. The kidney pelvis is surrounded by a thick wall of firm connective tissue in which are seen connective tissue cells with long, thin, darkly staining nuclei, whose long axes run parallel to the line of the fibres. Inside of this area there is a rather broad mesh-like layer of connective tissue in which there are large numbers of cellular fibroblasts and lymphocytes. There are also seen in this same area a great many eosinophile cells. The blood-vessels show rather thick walls, particularly the adventitial layers, and they are also congested. The straight tubules are atrophic and surrounded by connective tissue. The coiled tubules have disappeared in one portion of the section and are replaced by cellular connective tissue. Where they are present, their basement membranes are thickened by cellular growth. They show some cloudy swelling. The glomeruli are widely separated and greatly reduced in number. They show a rather thick capsule, and the glomerular coils have a marked increase in the number of endothelial cells. The interstitial connective tissue is everywhere increased and the picture has become, in the strict pathological sense, a chronic interstitial nephritis.

Ureter: The wall of the thinned-out ureter is lined by transitional epithelium which is very markedly diminished in thickness compared with the normal. The mucosa is composed of squamous cells, arranged in two layers, which are attached to a basement membrane. The submucosa seems to be thinned out in most places, and the muscular bundles are stretched out so that their long axis is in every case parallel with the axis of the ureter itself. There is a slight increase in connective tissue which is disposed between the stretched-out muscular elements, and there are many newly developed fibroblasts seen.

The bladder wall is tremendously thickened, and on microscopical

examination it is observed that there is an increase in all the elements composing the structure. The muscle bundles are much increased in size, and not only do there seem to be a great many more than the usual number of muscle fibres in the bundles, but the fibres themselves appear to be larger than the ordinary smooth fibres of the bladder. The connective tissue elements are also greatly increased. The intramuscular connective tissue bundles are large, and scattered here and there very frequently throughout the muscular bundles at their borders are seen collections of recently developed fibroblasts in nests or groups. There is a very abundant blood supply. The mucous membrane is of the usual transitional type. It is thickened as is the submucosa.

Urethra: The dilated prostatic urethra is lined with epithelium of the transitional type, resting upon a felt-work of fibro-elastic bundles under which is a thickened submucosa. The verumontanum is interesting, in that, by the use of Van Giesen's stain, it is observed that while there is a sprinkling of muscular elements throughout, it is at its lower portion made up mostly of connective tissue fibres. It is quite vascular throughout its entire length, the vessels being rather more numerous and considerably larger than is usually the case. Cross-section at the point of obstruction shows a very small slit-like lumen lined with stratified epithelium, resting on a felt-like base under which is a very thick submucosa. Surrounding this there is an exceedingly extensive area rather densely arranged made up of connective tissue and smooth muscle fibres. There are a number of dilated vessels quite thickly scattered throughout this tissue.

Discussion.—The case here described is very similar to the one described by Knox and Sprunt,[2] but considerably more pronounced. These writers made a very exhaustive study of the literature up to the time their article appeared, and found that while no reports on the subject were recorded in the American literature, a great many almost identical cases had been described in foreign journals: Wilckens,[3] Lederer,[4] Tolmatschew,[5] Bednar,[6] Godart,[7] Budd,[8] Velpeau,[9] Schlagenhaufer,[10] Commandeur,[11] Bonnet,[12] Teboul,[13] Fuchs,[14] Porak,[15] Lindeman,[16] and Picard [17] being among the number who had written on this subject. Bazy [18] observed six cases of congenital stricture in the region of the pars membranacea or pars bulbosa, in men and older boys, and emphasized the importance of congenital strictures even in adults. Ebert [19] punctured the obstructing membrane in a similar clinical case and discharged his patient as cured. Englisch [20] collected a large number of cases of congenital narrowing of the male urethra.

Recently, JORDAN [21] has published a case in which he found a stricture of the prostatic portion of the urethra in a boy about one year old, which measured one-quarter of an inch in length.

RIEDEL [22] has recently described nineteen cases of congenital stricture of the urethra that were treated in his clinic. Six of the nineteen died, a seventh

developed tuberculosis behind the narrowed portion of the urethra, but recovered. He considers it a relatively harmless developmental defect which only becomes serious when there is a considerable cutting off of the urethra, or when it is narrowed throughout its entire length. Dr. Riedel classified his cases as follows:

1. Contraction of the urethra behind the membranous portion.
2. Circumscribed area in front of the scrotum or an extensive network stricture at the perineum.
3. Stricture of the bulbous urethra.
4. Stenosis in front of external orifice and in perineum.

HENRY MORRIS[23] quotes a number of cases of congenital obstruction observed by himself and other European surgeons. Lamotte relieved a case, in which a thin membrane situated at the vesical orifice of the urethra had caused hydronephrosis, by passing a sound.

Howship (*Treatise on Diseases of Urinary Organs*) has described three cases of imperforate urethra and Dr. Kennedy (*Dublin Journ. Med. Sciences*) exhibited to the Dublin Pathological Society the body of a new-born infant with enormous distention of the urinary organs, due to obstruction of the urethra. Dr. Morris describes a case in which the kidneys, ureters and bladder were greatly distended, the only ascertainable cause being a small cyst in the mucous lining of the membranous urethra. This cyst did not quite fill the lumen of the urethra, but, being situated on the floor of the passage, it formed an effective barrier to the outflow of urine.

DR. HUGH H. YOUNG,[24] of Baltimore, has had several cases of congenital obstruction at the posterior urethra, two of which were similar to the case here described and both relieved by passing sounds. Another one was of a recent type, at the vesical orifice, and another a very peculiar double urethra, both of which he operated upon through the suprapubic route.

This case seems to be an anomaly of the development of the Wolffian and Müllerian ducts rather than a defect in the urethra itself. These structures, which in the male become the ejaculatory ducts, and the utriculus prostaticus, enter the prostate near its base and course in an oblique direction through that organ until they approach the urethra, at which point they turn and for a short distance run parallel with its axis, finally opening into its lumen. During their passage through the prostate, their musculature is bound together by a very definite, firm sheath of connective tissue. On their approach to the urethra, they push the floor of that structure up into a mound, forming the verumontanum, and still are separate from all other structures, their tissues being superimposed upon those of the urethra. Ordinarily, immediately below the openings of the ejaculatory ducts and utricle, the tissues surrounding them become distributed among the fibres on the floor of the urethra. Thus the verumontanum gradually becomes smaller and smaller until just below the apex of the prostate it disappears completely, spreading out laterally in the form of little bands which dis-

appear on the floor or walls of the urethra. In the specimen under consideration, although a number of fibres are disposed in the usual manner, the majority of them continue downward on the floor of the urethra. At the point where the verumontanum usually disappears, these fibres attach themselves to the entire circumference of the urethra, with the exception of a small portion of the floor to the left of the median line, thus producing an almost complete blocking of the urinary passage.

The dilatation of the urinary tract must have begun in this case as soon as any considerable amount of urine was secreted by the kidneys. On account of the obstruction to urinary outflow, the bladder undoubtedly filled up, and its continual contraction, in an attempt to empty itself, caused the enormous thickening of its wall and, by hydrostatic pressure of long standing, dilated the vesical sphincter to such an extent that it became absolutely ineffectual, a small ridge being left to mark its site. The posterior urethra thus became cone-shaped and directly continuous with the bladder lumen. The fact that the bladder wall has developed such an extensive hypertrophy instead of undergoing a thinning out process with separation of muscular bundles and enormous dilatation of the bladder lumen supports strongly the idea that the process has been very gradual and of long duration. By continually working against a gradually increasing pressure, the muscular fibres have become larger, more numerous and more compactly arranged so that the bladder lumen is only slightly if at all dilated. By the great thickening of its wall, the bladder has held the ureters in their passage through almost down to their normal size, although even under these conditions they are somewhat dilated in their course through the bladder wall and their sphincteric control has been totally or partially lost. The ureters and the pelves of the kidneys apparently do not respond by an extensive thickening, comparatively speaking, or at least if they do so, at first the increase in pressure overcomes such thickening, and these structures, being architecturally less competent than the bladder and prostatic urethra, become enormously dilated and the ureters are markedly lengthened so that in order to be accommodated they are thrown into distorted curves and folds. No particular reason can be found for the fact that the right ureter and kidney pelvis are somewhat smaller in size, and the ureter a little longer than the left. Back pressure has also distorted the arrangement of the kidney tissue proper, so that the usual landmarks are not distinguishable, and microscopically that organ shows a chronic interstitial nephritis.

In hydronephrosis, we have the remarkable picture of an organ

secreting fluid under a gradually increasing pressure, produced by itself, which finally brings about its own destruction.

In children, a hypertrophied bladder wall with dilated ureters would seem to indicate a response on the part of the urinary tract to a gradually increasing obstruction of long standing, while a dilatation of the bladder, with thinning of its walls, is due to a more sudden and more complete obstruction to the urinary outflow.

It is rather difficult to explain why this particular anomaly should occur so frequently, as the point of obstruction in this and similar cases is above the junction of the pars membranacea and the pars bulbosa, which is the generally accepted [25] point of division between that part of the urethra which develops from the entoderm and that which is derived from the ectoderm. Felix [26] does not agree with the opinion expressed by Broman, but maintains that the entire urethra from the verumontanum outward is developed from the sinus urogenitalis and is of entodermal origin.

Undoubtedly, a great many of these cases have advanced to such a degree that there is very serious renal impairment at the time of birth, but since a number of them live for several years (Budd's patient was 16 years old; Lederer's 11 years; Wilcken's 2¼ years; Knox and Sprunt's case, 5 years), it seems most important to urge upon the medical profession that any children who show an urinary output below the normal, a dribbing of urine, abdominal masses in the kidney region, or other signs and symptoms pointing to a disturbance of the urinary organs, should have a thorough urethral exploration, and obstructions, such as the one here described, could be relieved by proper instrumental treatment.

This investigation was made in the Pathological Department of Bellevue Hospital.

The author expresses his most sincere thanks to Dr. Charles Norris, Director of Laboratory, and his assistants, whose generosity and assistance have made its publication possible, and to Dr. Hugh H. Young for many helpful suggestions. The illustrations were made in the photographic studio at Bellevue Hospital by Dr. Bryson, the Photographer to the Pathological Department.

BIBLIOGRAPHY

[1] Lowsley: Am. Jour. Anat., No. 3, July, 1912.

[2] Knox and Sprunt: American Journal of Diseases of Children, September, 1912, vol. 4, pp. 137–147.

[3] Wilckens: Zur Frage der Kongenitalen Stenosen der Männlichen Hornröhre. Ztschr. f. Urol., 1910, iv, 814.

[4] Lederer: Ueber eine Angeborene membranöse Verengerung der Pars prostatica Urethrae. Arch. f. path. Anat. u. Physiol., Berl., 1911, cciii, 240.
[5] Tolmatschew: Ein Fall von Semilunaren Klappen der Männlichen Hornröhre und Von Vergrösserter Vesicula Prostatica. Arch. f. path. Anat. u. Physiol. Berl., 1870, xlix, 348.
[6] Bednar: Quoted from Wilckens,[8] Lindeman,[16] *et al.*
[7] Godart: Valvule Anormale dans Le canal l'urètre. Bull. Soc. Anat. de Paris, 1854 xxix, 137.
[8] Budd: Quoted from Wilckens,[8] Lindeman,[16] *et al.*
[9] Velpeau: Traité d'anatomie Chirurgicale, Paris, 1826, ii, 297.
[10] Schlagenhaufer: Ein Beitrag zu den Angeborenen Klappenbildungen im Beireiche der Pars Prostatica urethrae. Wien. Klin. Wchnschr., 1896, No. 15, p. 268.
[11] Commandeur: Dilatation de l'Appareil urinaire Chez le foetus par retrécissement valvulaire Congénital de l'urètre. Lyon Méd., 1898, Mars. 13.
[12] Bonnet: Quoted from Wilckens.[8]
[13] Teboul: Quoted from Wilckens.[8]
[14] Fuchs: Zwei Fälle von Kongenitaler Hydronephrose. Inaug. Dis., Zurich, 1900.
[15] Porak: Quoted from Commandeur[11] and Lederer.[4]
[16] Lindeman: Casuistischer Beitrag Zur Frage der Angeborenen Klappenförmigen Verengerung der Pars Prostatica Urethrae. Inaug. Dis., Jena, 1904.
[17] Picard: Convulsions à forme eclamptique chez un homme. Gaz. méd. de Strassburg, 1855, No. 7, p. 259. Quoted from Lindeman.[16]
[18] Bazy: Rétrécissement Congénital de l' urètre chez l' homme. Presse Méd., 1903, p. 215.
[19] Ebert: Quoted from Segall, Ein Fall von Angeborener Hornröhrenverengerung. Inaug. Dis., Königsberg, 1890.
[20] Englisch: Ueber Angeborene Verengerungen der Männlichen Hornröhre. Folia Urologica, 1909, iv and v.
[21] Jordan: The Journal of The American Medical Association, July 26, 1913, vol. lxi, No. 4.
[22] Riedel: Archiv für Klinische Chirurgie., June 17, 1913, pp. 763–778.
[23] Surgical Diseases of Kidney and Ureter, vol. i.
[24] Young: Personal communication.
[25] Broman: Normale und abnorme-Entwicklung des Meuschen. Wiesbaden, 1911 (J. F. Bergmann), p. 476.
[26] Felix: Die Entwicklung der Harn und Geschlechts Organe. Handbuch der Entwicklungsgeschichte des Menschen (F. Keibel and F. P. Mall), ii, p. 732.